Long-Term Follow-Up of Persistent Truncus Arteriosus: Kuwait Experience

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Key Words

Persistent truncus arteriosus • Common arterial trunk • Homograft reconstruction • Conduit repair

Abstract

Objective: To evaluate the long-term results of patients in Kuwait who were operated for persistent truncus arteriosus (PTA). **Subjects and Methods:** The following data were collected for retrospective analysis from 24 medical records of consecutive patients with PTA in Kuwait between August 1993 and August 2009: demographics, morphology, management and outcome. Major associated abnormalities included interrupted aortic arch in 1 patient and abnormal coronary artery anatomy in 2. **Results:** Of the 24 patients, 16 underwent total intracardiac repair. The age at operation ranged from 15 days to 5 years (mean 166.19 ± 438.63 days) and weight ranged from 2.5 to 15 kg (mean 4.3 ± 3.01 kg). The right ventricle to pulmonary artery continuity was established with aortic homograft in 11, pulmonary homograft in 4 and by implantation of a Contegra conduit in 1 patient. Four patients had moderate truncal valve regurgitation requiring concomitant truncal valve repair. After a mean follow-up period of 81.81 ± 61.58 months (range 3–166) there was no death. Eight of the 16 (50%) patients underwent redo homograft operations. One patient who had concomitant truncal valve repair subsequently underwent aortic valve re-

placement. **Conclusion:** The data showed that complete repair of PTA in the neonatal and early infancy period was the treatment with the best potential for survival. The homograft remained one of the conduits of choice to establish continuity between the right ventricle and the pulmonary artery in spite of the high incidence of conduit redo operations.

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Introduction

Persistent truncus arteriosus (PTA) also known as common arterial trunk is best described as a congenitally malformed heart with normal atrial arrangement and concordant atrioventricular connections in which a solitary arterial trunk arises from the base of the heart and supplies the coronary, pulmonary and systemic arteries [1]. It accounts for 1.2–3% of all congenital heart malformations. This malformation is eventually fatal if not treated. The mean age of death is 2.5 months with 80% of affected children dead by 1 year of age [2]. The definitive treatment of PTA is surgery. The physiologic correction of PTA was described by McGoon et al. [3] in 1968. Since then, surgical correction during infancy has become possible. Here we report our long-term follow-up experience of patients with surgical repair of PTA.

Table 1. Demographic and clinical data of the study group

Case No.	Year of birth	Age at presentation	Age at operation	Sex	Weight at operation, kg	Van Praagh classification	Truncal valve leaflet	Truncal valve regurgitation	Clinical symptoms			Associated cardiac anomaly	Associated noncardiac anomaly	Outcome
									CHF	cyanosis	FTT			
1	1992	9 days	22 days	F	3	A1	tricuspid	mild	+	-	-	-	-	operated
2	1994	9 days	expired	F	-	A2	tricuspid	mild	+	-	-	ASD	-	expired
3	1994	45 days	68 days	F	3.1	A1	tricuspid	mild	+	-	+	-	-	operated
4	1995	13 days	15 days	M	2.7	A1	quadricuspid	mod.	+	-	-	-	-	operated
5	1995	40 days	60 days	F	3	A1	quadricuspid	mild	+	+	+	ASD	-	operated
6	1995	23 days	29 days	M	2.9	A2	tricuspid	mild	+	+	-	abnormal coronaries	-	operated
7	1995	5 months	5 months	F	4.8	A2	quadricuspid	mild	+	+	+	-	-	operated
8	1996	11 days	60 months	M	15	A3	tricuspid	mild	+	+	+	PDA	G6PD deficiency	operated
9	1996	12 days	21 days	M	3.4	A3	bicuspid and dysplastic	mod.	+	-	-	IAA type A, PDA	-	operated
10	1998	2 days	25 days	F	3.1	A1	quadricuspid	mild	+	-	-	-	-	operated
11	1998	4 days	22 days	M	2.6	A2	bicuspid and dysplastic	mild	+	-	-	-	-	operated
12	1998	7 days	1 month	F	3.5	A1	tricuspid	mod.	+	+	-	-	-	operated
13	1999	7 days	17 days	M	3.4	A1	tricuspid	mild	+	-	-	RAA, RPA stenosis	-	operated
14	2002	1 year	not operated	F	-	A1	tricuspid	mild	+	+	+	PDA	-	not operated
15	2004	1 day	not operated	F	-	A1	tricuspid	nil	+	+	-	-	-	not operated
16	2004	1 day	expired	M	-	A2	bicuspid and dysplastic	mild	+	+	-	RAA	-	expired
17	2005	24 days	28 days	F	2.5	A1	quadricuspid	mod.	+	-	-	-	hydronephrosis	operated
18	2006	1 day	expired	F	-	A4	tricuspid	nil	+	-	-	-	-	expired
19	2006	1 day	expired	M	-	A1	tricuspid	mild	+	+	-	-	-	expired
20	2006	55 days	4 months	M	4.9	A1	tricuspid	mild	+	-	-	mild MVP	-	operated
21	2007	1 day	expired	M	-	A1	quadricuspid	mild	-	-	-	hypoplastic PA	-	expired
22	2008	7 days	expired	F	-	A1	quadricuspid	mild	+	-	-	ASD	-	expired
23	2008	67 days	72 days	F	5	A1	tricuspid	mild	+	+	+	RAA, coronary abnormality	-	operated
24	2008	2 months	6 months	M	5.4	A1	tricuspid	mild	+	+	+	-	G6PD deficiency	operated

CHF = Congestive heart failure; FTT = failure to thrive; ASD = atrial septal defect; mod. = moderate; PDA = patent ductus arteriosus; RAA = right aortic arch; RPA = right pulmonary artery; MVP = mitral valve prolapse; PA = pulmonary artery.

Subjects and Methods

The medical records of a group of 24 consecutive infants from Kuwait, who underwent corrective repair of PTA from August 1993 to June 2009, were reviewed (table 1). All patients were diagnosed by two-dimensional cross-sectional and colour Doppler echocardiography. In 7 patients preoperative cardiac catheterization was performed for better characterization of the diagnosis. The patients were classified according to Van Praagh classification (fig. 1): type A1 = 16 patients; type A2 = 5 patients; type A3 = 2 patients and type A4 = 1 patient. The truncal valve leaflets were bicuspid and dysplastic in 3, tricuspid in 14 and quadricus-

pid in 7 patients. Mild truncal valve regurgitation was present in 18 and moderate regurgitation in 4 patients. Moderate truncal valve regurgitation was associated with 1 patient with bicuspid dysplastic valve, 1 with tricuspid valve and 2 with quadricuspid valve. Coronary abnormalities were observed in 2 patients in our series. The mean age at presentation was 38.13 ± 77.31 days (range 1–365). Six patients died while waiting for surgery, 1 due to extreme prematurity and the others due to fulminant sepsis, necrotizing enterocolitis and disseminated intravascular coagulation. The parents of 2 infants did not accept surgery for their babies and returned to their native countries. The ventricular septal defect was closed with a patch in all of them through a right ven-

Fig. 1. Van Praagh classification of PTA (courtesy: Children's Hospital, Boston, Mass., USA).

tricolotomy and then a homograft was anastomosed first to the main pulmonary artery or its branches and then to the right ventriculotomy. The assessment of the functional result and capacity was performed in surviving patients by clinical, radiological, electrocardiographic and cross-sectional echocardiographic and colour flow Doppler examinations.

Results

Of the 24 patients, 16 (66.66%) underwent surgical repair. The details of results are described in tables 1 and 2. The mean age at operation was 166.19 ± 438.63 days (range 15–1,825). Nine patients were operated before 1 month of age, 4 between 1 and 3 months, 1 each at 5, 6 and 60 months, respectively. One patient was diagnosed initially as pulmonary atresia with ventricular septal defect and a right modified Blalock-Taussig shunt was performed at 18 months of age. Later at 5 years of age, it was recognized during surgery as type A3 PTA and corrective surgery was done. The mean weight at operation was 4.3 ± 3.01 kg (range 2.5–15). The mean size of the original conduit used was 11.87 ± 1.93 mm (range 10–18). Of the 16 patients, 11 (68.7%) had an aortic homograft, while 4 (25%) had a pulmonary homograft. One (6.25%) patient had implantation of a Contegra conduit between the right ventricle and pulmonary artery. One patient with associated interrupted aortic arch (IAA) underwent simultaneous aortic arch repair.

There was no death in the postoperative period. These 16 patients had a mean follow-up period of 81.81 ± 61.58 months (range 3–166). Eight patients (50%) underwent redo homograft replacement at 10, 24, 27, 50, 105, 143, 144 and 166 months (mean 83.63 ± 62.9 months; range 10–166) after the initial operation. Two (25%) had a pulmonary homograft in the first operation while 6 (75%) had aortic homograft. Seven of them had moderate to severe homograft stenosis with a gradient above 50 mm Hg pri-

or to replacement. Four patients had moderate or more truncal valve regurgitation and underwent concomitant valve repair during the initial corrective surgery. The regurgitation was persistently severe in 1 patient and this child had his truncal valve replaced with St. Jude Medical valve prosthesis at 9 years of age. Seven of the remaining 8 patients who survived with the first homograft had mild homograft stenosis and 1 had no stenosis at last follow-up. Amongst the 16 patients who had initial surgery, mild homograft regurgitation was found in 11 (68.75%) and moderate regurgitation in 2 (12.5%). One patient (6.25%) had moderate aortic valve (previous truncal valve) regurgitation while 8 (50%) patients had mild aortic regurgitation and 3 (18.75%) had mild aortic stenosis. The left ventricular systolic function was normal (ejection fraction more than 60%) in all those followed up. Thirteen of the 16 (81.25%) patients who were followed up had symptoms of NYHA class 1, 2 (12.5%) of class 2 and 1 (6.25%) of class 3. All school-age children attend normal school.

Discussion

The follow-up of patients undergoing physiologic correction showed that a high percentage suffered from pulmonary vascular disease, which occurs at a fairly young age in the majority of patients with PTA [4]. A policy of early complete correction of symptomatic infants with PTA was adopted in 1974 [5]. Severe truncal valve regurgitation, IAA, coronary artery anomalies and age at repair greater than 100 days were important risk factors for perioperative death [6]. The 4 children in this study who were more than 100 days of age at the time of initial corrective surgery, 2 with coronary artery abnormalities and the only child with associated type A IAA, did well on follow-up. One-stage repair of both PTA and IAA is the optimal management [7].

Table 2. Findings of postoperative follow-up

Case No.	Conduit and size used	Concomitant heart surgery	Associated morbidity	Conduit replacement	Time after initial surgery, months	Follow-up period, months	Last NYHA status
1	12-mm pulmonary homograft	–	mod. HG stenosis, mild AR	21-mm aortic homograft	144	159	1
3	12-mm aortic homograft	truncal valve repair	mod. HG stenosis, Mod. AR	21-mm aortic homograft	24	144	1 and after AV replacement
4	10-mm pulmonary homograft	truncal valve repair	temporary CHB, mild AR, mild HG Reg	–	–	3	2 and lost to follow-up
5	11-mm aortic homograft	–	mod. HG stenosis, mild HG Reg	17-mm aortic homograft	50	144	1
6	12-mm aortic homograft	–	mild AR, mild HG stenosis, mild HG Reg	–	–	3	1 and lost to follow-up
7	10-mm pulmonary homograft	–	mod. HG stenosis, mild HG Reg, mild AR	18-mm Hancock valved conduit	143	146	1
8	18-mm aortic homograft at 60 months	removing of previous BT shunt at 18 months	mod. HG stenosis, mild HG Reg	21-mm pulmonary homograft	166	166	1
9	11-mm aortic homograft	truncal valve repair + aortic arch repair	SVT, sepsis, mild HG stenosis and Reg	–	–	42	1
10	11-mm aortic homograft	–	mod. HG stenosis, mild AR, CHF	21-mm pulmonary homograft	105	108	3
11	10-mm aortic homograft	–	SVT, mild AR, mild HG stenosis, mod. HG Reg	11-mm porcine valved conduit	10	75	2
12	12-mm aortic homograft	truncal valve repair	mild HG stenosis, mild AR, mod. HG Reg	–	–	132	1
13	12-mm aortic homograft	–	mod. HG stenosis, mild HG Reg, mild AR	21-mm pulmonary homograft	27	88	1
17	11-mm aortic homograft	–	mild HG stenosis, mild HG Reg, mild AS	–	–	8	1
20	12-mm aortic homograft	–	mild HG Reg, mild HG stenosis	–	–	58	1
23	12-mm pulmonary homograft	–	mild HG stenosis, mild HG Reg, mild AS	–	–	25	1
24	14-mm Contegra conduit	–	mild HG stenosis, mild HG Reg, mild AS	–	–	8	1

mod. = Moderate; HG = homograft; AR = aortic regurgitation; AV = aortic valve; CHB = complete heart block; Reg = regurgitation; BT = Blalock-Taussig; CHF = congestive heart failure; SVT = supraventricular tachycardia; AS = aortic stenosis.

Only 1 patient in this study had implantation of Contegra conduit consistent with the recommendation of Gober et al. [8] not to routinely use Contegra valved conduit for reconstruction of the right ventricular outflow tract because of an unpredictable incidence of supra-valvar stenosis during midterm results. However, the child was still doing well with the initial surgery. Cryopreserved homograft conduits are generally preferred for the initial repair due to their ease of implantation and low incidence of conduit stenosis [9]. The majority of the patients (93.75%) in this study had cryopreserved homograft conduits for the initial repair. Sinzobahamvya et al. [10] had made a similar observation in their study of 35 patients. The decision regarding the type of right ventricle to pulmonary artery connection surgery was at the

discretion of the attending surgeon. Fifty percent of the patients of this study underwent conduit reoperations in a mean period of 83.63 ± 62.9 months (range 10–166). In spite of the reported high (50%) incidence of conduit re-interventions, the excellent result of no late mortality may be due to the predominant use of homograft. Conduit replacement is eventually necessary in all cases because of homograft valvular stenosis due to shrinkage, distal anastomosis stenosis, conduit calcification, proximal hood aneurysm and somatic outgrowth [11]. Rajasinghe et al. [12] and Tlaskal et al. [13] in their studies of long-term follow-up of PTA after surgical correction reported a similar high incidence of conduit reoperations. In PTA type A1 and A2, Barbero-Marcial et al. [14] and Nemoto et al. [15] have described alternative techniques of right

ventricle to pulmonary artery continuity established by direct anastomosis. This has potential for the right ventricular outflow tract growth and had a low incidence of surgical reinterventions of right ventricular outflow tract. Recent studies by Raisky et al. [16] and Xu and Shen [17] concluded that repair of PTA by direct anastomosis of right ventricle to pulmonary artery continuity does not increase mortality and morbidity but decreases the need for reinterventions. The 4 patients who had moderate truncal valve regurgitation in our study had their truncal valve repaired in the primary stage. Kaza et al. [18] in one of the largest series of truncal valve repair emphasized that truncal valve repair should remain the primary option. One of the 4 had to undergo valve replacement 9 years later due to persistent severe regurgitation.

The limitations in this study were the small number of the study size and the preferred surgical choice of using homograft. Further studies of a larger study size are needed to draw significant conclusions.

Conclusion

Our data showed that complete repair of PTA in the neonatal and early infancy period was the treatment with the best potential for survival. Homograft remains one of the conduits of choice to establish continuity between the right ventricle and pulmonary artery in spite of the high incidence of conduit redo operations.

References

- Schofield DE, Anderson RH: Common arterial trunk with pulmonary atresia. *Int J Cardiol* 1988;20:290–294.
- Yoskizato T, Julsrud PR: Truncus arteriosus revisited: an angiographic demonstration. *Pediatr Cardiol* 1990;11:36–40.
- McGoon DC, Rastelle GC, Ongley PA: An operation for the correction of truncus arteriosus. *JAMA* 1968;205:69.
- McGoon DC, Wallace RB, Danielson GK: The Rastelle operation: its indication and results. *J Thorac Cardiovasc Surg* 1973;65:865.
- Singh AK, De Leval MR, Pincott JR, Stark J: Pulmonary artery banding for truncus arteriosus in the first year of life. *Circulation* 1976;54:17–19.
- Haniev FL, Heinemann NK, Jonas RA, Mayer JE, Cook N, Wessel DL, Castaneda AR: Repair of truncus arteriosus in the neonate. *J Thorac Cardiovasc Surg* 1993;105:1047–1056.
- Kanstantinov IE, Karamlov T, Blackstone EH, Mosca RS, Lofland GK, Caldarone CA, Williams WG, Mackie AS, McGrindle BW: Truncus arteriosus associated with interrupted aortic arch in 50 neonates: a Congenital Heart Surgeons Society Study. *Ann Thorac Surg* 2006;81:222–223.
- Gober V, Berdt P, Pavlovic M, Pfamatter JP, Carrel TP: Adverse midterm outcome following RVOT reconstruction using the Contegra valved bovine jugular vein. *Ann Thorac Surg* 2005;79:625–631.
- Clarke DR, Campbell DN, Bishop DA: Homograft for congenital heart disease; in Kron IL (ed): *Cardiac Surgery: State of the Art Reviews*. Philadelphia, Hanley & Belfus, 1989, vol 3, pp 363–379.
- Sinzobahamvya N, Boscheinen M, Blaschczoc HC, Kallenberg R, Photiadis J, Haun C, Hraska V, Asfour B: Survival and intervention after neonatal repair of truncus arteriosus with valved conduit. *Eur J Cardiothorac Surg* 2008;34:732–737.
- Bove EL, Beekman RH, Snider AR, Callow LB, Underbill DJ, Rocchini AP, Dick M, Rosenthal A: Repair of truncus arteriosus in the neonate and young infant. *Ann Thorac Surg* 1989;47:499–506.
- Rajasinghe HA, McElhinney DB, Reddy VM, Mora BN, Hanley FL: Long-term follow-up of truncus arteriosus repaired in infancy: a twenty-year experience. *J Thorac Cardiovasc Surg* 1997;113:869–879.
- Tlaskal T, Chaloupecky V, Hucin B, Gebauer R, Krupickova S, Reich O, Skovranek J, Tax P: Long-term results after correction of persistent truncus arteriosus in 83 patients. *Eur J Cardiothorac Surg* 2010;37:1278–1284.
- Barbero-Marcial M, Riso A, Attk E, Jatene A: A technique for correction of truncus arteriosus types I and II without extra cardiac conduits. *J Thorac Cardiovasc Surg* 1990;99:364–369.
- Nemoto S, Ozawa H, Sasaki T, Katsumata T, Kishi K, Okumura K, Mori Y: Repair of persistent truncus arteriosus without a conduit: sleeve resection of the pulmonary trunk from the aorta and direct right ventricle-pulmonary artery anastomosis. *Eur J Cardiothorac Surg* 2011;13:278–284.
- Raisky O, Ali WB, Bajolle F, Marini D, Metton O, Bonnet D, Sidi D, Vouhe PR: Common arterial trunk repair: with conduit or without? *Eur J Cardiothorac Surg* 2009;36:675–682.
- Xu ZW, Shen J: Repair of truncus arteriosus: choice of right ventricle outflow reconstruction. *J Card Surg* 2010;25:724–729.
- Kaza AK, Burch PT, Pinto N, Minich LL, Tani LY, Hawkins JA: Durability of truncal valve repair. *Ann Thorac Surg* 2010;90:1307–1312.